

Dual Clinical Presentation of a Birthmark: A Quiz

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A man in his early 30s presented with a longstanding birthmark on the left thigh that was first noticed at age 12. As a teenager, the lesion was treated with a CO₂ laser at an outside institution; however, the lesion continued to slowly enlarge over the past several years. The patient reported recurrent irritation and swelling from a construction safety harness with occasional bleeding and frequent bruising. Recently, the area became inflamed, prompting an ER visit, where the patient was evaluated for cellulitis, underwent a normal ultrasound and was placed on cephalexin 500 mg twice daily for 10 days.

Clinically, the lesion consisted of clusters of dark red to blue-black papules coalescing into a large plaque with adjacent areas of purpura (Fig. 1A). A punch biopsy showed superficially located dilated vascular spaces

congested with erythrocytes with chronic inflammation (Fig. 2A).

On follow-up 2 months later, the large plaque had changed appearance, showing numerous grouped translucent pink vesicles forming a large plaque on the left thigh, with a few satellite haemorrhagic vesicles (Fig. 1B). Repeat punch biopsy revealed dilated lymphatic channels lined by flattened endothelial cells with thin fibrous septa and sparse lymphocytes (Fig. 2B).

What is your diagnosis.

- 1: Angioma serpiginosum.
- 2: Angiokeratoma circumscriptum.
- 3: Microcystic Lymphatic Malformation.
- 4: Lymphangiectasia.

See next page for answer.

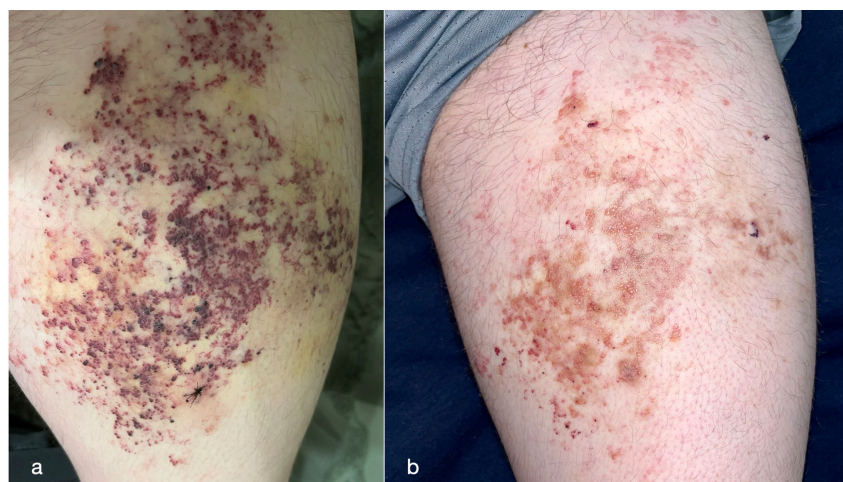


Fig. 1. (A) Initial visit after safety harness-related trauma. (B) Follow-up visit, trauma to birthmark resolved.

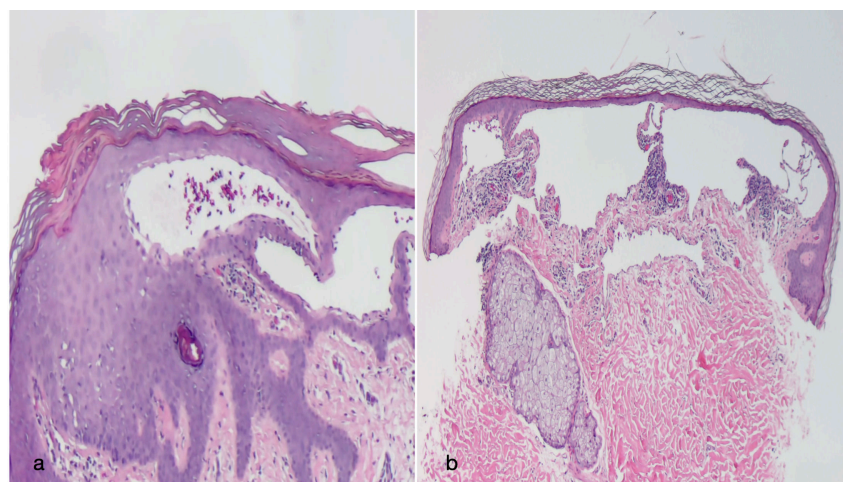


Fig. 2 (A) Initial biopsy: superficially located dilated vascular spaces filled with erythrocytes. (B) Follow-up biopsy: dilated lymphatic channels lined by flattened endothelial cells with thin fibrous septa.

ANSWERS TO QUIZ

Dual Clinical Presentation of a Birthmark: A Commentary

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Diagnosis: Microcystic Lymphatic Malformation (MiLM).

Lymphatic malformations are uncommon hamartomatous anomalies of the lymphatic system that often involve the skin and subcutaneous tissue (1, 2). Historically, lymphatic malformations were classified by depth into a sup form, called lymphangioma circumscriptum, and deep forms, referred to as cavernous lymphangioma and cystic hygroma (2, 3). They are now organized by the International Society for the Study of Vascular Anomalies (ISSVA) classification, which divides vascular anomalies into tumours and malformations. Under this system, lymphatic malformations are categorized as common (cystic), channel-type, complex or primary lymphedema, with common (cystic) lesions further subdivided by cyst size into microcystic (<1 cm) or macrocystic (>1 cm), with many lesions showing combined features. Microcystic Lymphatic Malformation (MiLM) is the most common variant, typically presenting as a collection of superficially dilated lymphatic vessels measuring <1 cm in diameter (4–6).

Lymphatic malformations may be congenital or acquired. Congenital lymphatic malformations are thought to occur due to deep lymphatic channels that remain abnormally connected to superficial dermal lymphatics but fail to join the normal drainage system, leading to fluid accumulation and dilation of the superficial channels (7). Acquired lymphatic malformation, such as lymphangiectasia, occur from secondary dilation of previously normal lymphatics due to obstruction and have been described in areas of chronic lymphedema as well as following infections, radiation therapy and morphea (1–3). Our patient first noticed the lesion at age 12 but denied preceding infection or trauma, suggesting it was likely present in a more subtle form from an earlier age. More recently, a construction safety harness worn at work likely aggravated the skin surface and may have caused transient external compression proximal to the lesion, further obstructing lymphatic drainage of the left thigh and contributing to the initial presentation.

MiLM typically presents as clusters of multiple translucent or haemorrhagic thin-walled vesicular papules that resemble “frog spawn.” They may appear darkened when blood is present (3). These lesions are usually asymptomatic but may be complicated by swelling, bruising, haemorrhage, ulceration or recurrent cellulitis, leading to changes in clinical appearance and possible misdiagnosis, as initially occurred in our patient (1, 3). Common sites of involvement include the proximal extremities, axillary folds, shoulders, neck, trunk, buccal mucosa and perineum.

The differential diagnosis includes herpes zoster, molluscum contagiosum, viral warts, angiokeratomas, haemangiomas, epidermal nevi, leiomyomas and angiosarcoma (3, 4). In our case, angioma serpiginosum, angiokeratoma circumscriptum and lymphangiectasia were considered. Angioma serpiginosum consists of punctate clustered or serpiginous nonblanchable erythematous macules with histology showing dilated capillaries in the papillary dermis. Angiokeratoma circumscriptum presents as clustered dark red to violaceous keratotic papules and shows papillary dermal vascular ectasia with overlying hyperkeratosis and papillomatosis. Lymphangiectasia is similar histologically to MiLM but acquired from lymphatic obstruction or injury.

Histopathology shows thin-walled, dilated lymphatic vessels in the superficial dermis, occasionally extending into the reticular dermis or subcutis. The vessels are lined by endothelial cells and may contain eosinophilic material or red blood cells. The overlying epidermis is often acanthotic or hyperkeratotic (3).

Treatment of MiLM is challenging. When feasible, surgical excision is a definitive treatment option for localized lesions, with recurrence rates up to 23%, although wound-healing complications and worsening may develop from further disruption of lymphatic channels. Less invasive options are often used to control symptoms, but recurrence is common. CO2 laser, electrocautery and radiofrequency ablation may be used but can be associated with pain or scarring. Topical imiquimod has shown benefit, with local irritation common. Intralesional sclerotherapy can potentially target deeper involvement, although swelling, pain or inflammation may occur. Pulsed dye laser may reduce erythema and the vascular aspect of the malformation, with less downtime and minimal scarring (3, 8, 9).

T2-weighted MRI is useful for evaluating the depth and extent of lymphatic malformations. For deeper or more extensive lesions, percutaneous bleomycin sclerotherapy and rapamycin have shown benefit (10). Our patient has improved with 2 sessions of 595 nm pulsed dye laser using a 5 mm spot size, 8–10 J/cm² fluence and 1.5 ms pulse duration, and we plan to continue periodic laser treatments to manage the lesion effectively. Should laser therapy fail to achieve a durable response, the patient has agreed to MRI to evaluate for deeper involvement and guide further management.

REFERENCES

- Kandula S, Mark L, Warren S. Acquired lymphangioma circumscriptum. J Am Acad Dermatol 2012; 66: AB41. <https://doi.org/10.1016/j.jaad.2011.11.181>
- Tan SK, Tay YK. Lymphangioma circumscriptum. N Engl J Med 2012; 366: 1724–1724. <https://doi.org/10.1056/NEJM1101005>
- Miceli A, Stewart KM. Lymphangioma. In: StatPearls. StatPearls Publishing; 2023.
- McCormack L, Jones K, Huang JT. Micro- and macrocystic lymphatic malformation. J Pediatr 2020; 219: 275–276. <https://doi.org/10.1016/j.jpeds.2019.12.004>

5. Edminister JR, Zarbo A, Seale L, Friedman BJ, Shwayder T. Microcystic lymphatic malformation presenting as firm, skin-colored papules of the lips. *Pediatr Dermatol* 2022; 39: 992–994. <https://doi.org/10.1111/pde.15060>
6. International Society for the Study of Vascular Anomalies. ISSVA classification for vascular anomalies. 2018. [cited 2026 June 14]. Available from: <http://issva.org/classification>
7. Whimster IW. The pathology of lymphangioma circumscriptum. *Br J Dermatol* 1976; 94: 473–486. <https://doi.org/10.1111/j.1365-2133.1976.tb05134.x>
8. Puri N. Treatment options of lymphangioma circumscriptum. *Indian Dermatol Online J* 2015; 6: 293–294.
9. Bikowski JB, Dumont AMG. Lymphangioma circumscriptum: treatment with hypertonic saline sclerotherapy. *J Am Acad Dermatol* 2005; 53: 442–444. <https://doi.org/10.1016/j.jaad.2005.04.086>
10. Petkova M, Ferby I, Mäkinen T. Lymphatic malformations: mechanistic insights and evolving therapeutic frontiers. *J Clin Invest* 2024; 134: e172844. <https://doi.org/10.1172/JCI172844>